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Case Report

Twiddler's syndrome detected by patient's complaint of implantable cardioverter-defibrillator rotation in the subcutaneous pocket

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ABSTRACT

Twiddler's syndrome presents as a rare implantable-cardioverter defibrillator (ICD) malfunction that is potentially fatal. The following case is that of a 79-year-old woman, who was implanted with an ICD and experienced Twiddler's syndrome. Five months after the implantation, the patient complained that the generator sometimes rotated in the subcutaneous pocket. Fluoroscopy showed that the generator had rotated and the leads were twisted, which is compatible with Twiddler's syndrome. Although all the leads were functioning normally, a procedure to repair the twisted leads was performed. Twiddler's syndrome is usually detected due to a serious device malfunction; however, this case study suggests that it can be detected prior to ICD malfunction by the patient's complaint.

1. Introduction

Twiddler's syndrome has been described as a rare complication of pacemakers and implantable cardioverter-defibrillators (ICD), which typically leads to device malfunction [1–15]. We present a case of Twiddler's syndrome that was detected by the patient's complaint of generator rotations in the subcutaneous pocket before any ICD malfunction occurred.

2. Case report

A 79 year-old woman (height: 154.0 cm, weight: 60.6 kg) with coronary artery disease received an ICD (Current DR, model CD2211-36Q, St. Jude Medical, Sylmar, CA, USA) due to hemodynamically unstable sustained ventricular tachycardia. An active fixation ventricular lead (Durata, model 2120Q; St. Jude Medical, Sylmar, CA, USA) was placed in the ventricular mid-septum and an active fixation atrial lead (Fineline, 4473-52; Boston Scientific Corp., St. Paul, MN, USA) was implanted in the right atrial anterior wall by using the cephalic vein cutdown technique (Fig. 1). Acceptable lead impedance and capture/sensing thresholds were obtained at implantation (Table 1). Five months later, the patient reported that the generator sometimes rotated in the subcutaneous pocket when she turned over in bed. Fluoroscopy showed that the generator had rotated and the leads were twisted, indicating Twiddler's syndrome (Fig. 2). Telemetry data did not demonstrate any lead malfunction, and no delivery of inappropriate shocks was experienced. Only a slight increase in the capture threshold of the shock lead was observed (Table 1). Therefore, the leads were untwisted and fixation of the generator to the pectoral muscle was performed (Fig. 3).

3. Discussion

Twiddler's syndrome has been known to be an uncommon cause of malfunction in pacemakers, ICDs, and cardiac resynchronization therapy devices [1–15]. The prevalence of this syndrome was reported to be 0.07% in cases with a pacemaker [2,7]. This syndrome occurs if the patient consciously or unconsciously twiddles the implanted generator in the subcutaneous pocket [4]. Twiddling of the pulse generator can result in twisting of the leads within the pocket; this usually results in dislocation or fracture of the leads, which causes loss of capture and abnormal

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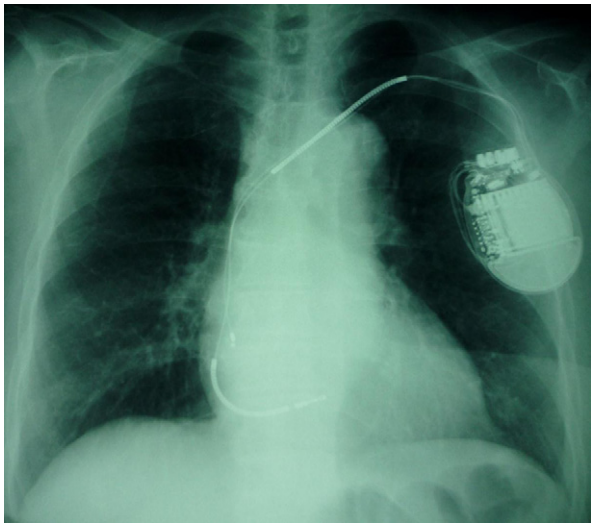

Fig. 1. Chest roentgenography performed at the time of ICD implantation.

Table 1
Impedance, capture threshold, and sensing just after implantation of the device and at the time of diagnosis of Twiddler's syndrome.

	Just after implantation	At diagnosis of Twiddler's syndrome
Atrial Lead		
Impedance (Ω)	390	340
Capture threshold (V/0.5 ms)	0.75	0.75
Sensing (mV)	3.1	2.3
Ventricular Lead		
Impedance (Ω)	290	310
Capture threshold (V/0.5 ms)	1.75	2.0
Sensing (mV)	4.9	4.6

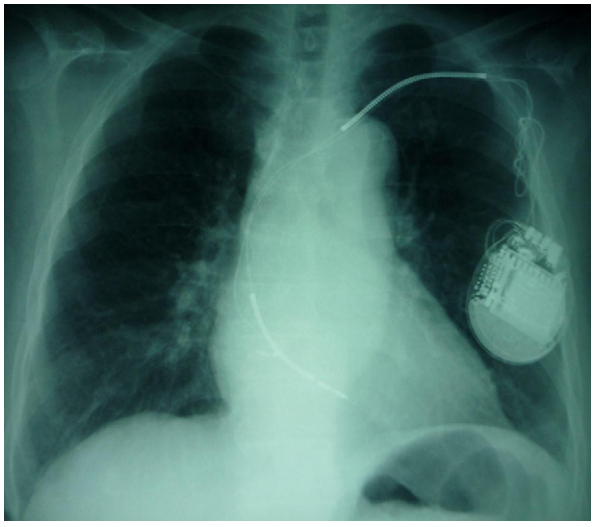

Fig. 2. Chest roentgenography performed at the time of diagnosis with Twiddler's syndrome, 5 months after the implantation. The ICD has turned over and the leads were twisted in the pectoral pocket.

sensing [1–15]. In these cases, the electrocardiogram shows a failure to capture and the chest radiography reveals the dislodged and twisted leads. Hypoperfusion symptoms, such as fatigue and syncope, are observed in pacing-dependent patients. In a few reports, the dislodged lead caused ipsilateral phrenic nerve stimulation leading to the sensation of diaphragmatic and abdominal pulsations

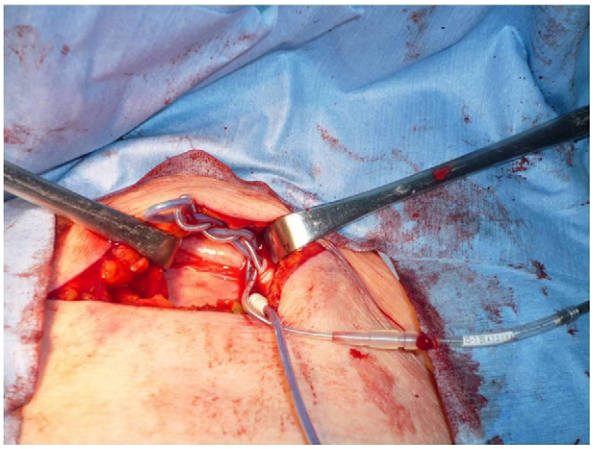

Fig. 3. Intraoperative image showing the twisting of the leads caused by several rotations of the defibrillator around the longitudinal axis.

[6,11,14]. Twiddler's syndrome is more serious and potentially life threatening in cases with a defibrillator because treatment of ventricular arrhythmias would be disabled and inappropriate shocks could be delivered due to over sensing [10,12,13].

According to most published reports, patients with Twiddler's syndrome are usually diagnosed due to some type of sudden device malfunction [1–4,6–15]. Early detection of this condition can also be achieved with a home monitoring system [8]. The case presented is a rare one in which Twiddler's syndrome was detected before device malfunction occurred. According to published reports, the patient's unconscious twiddling of the generator is typically the cause of this syndrome. However, the patient in the case presented did not have any mental disorders and denied having rotated the generator herself. The patient recognized and reported that the generator sometimes rotated in the subcutaneous pocket, which was helpful for the early diagnosis. In addition, lead dislodgment may have been avoided by using active fixation leads.

Twiddler's syndrome has been described as presenting complications in cases where the subcutaneous tissues are lax, the generator is untethered in its pocket, or the size of the pocket exceeds that of the generator. Patients who are most at risk of this condition include middle-aged, obese women, and patients with mental disorders such as dementia [3]. Lax subcutaneous tissue was the most plausible risk factor in the case presented. If several risk factors for Twiddler's syndrome, such as obesity, excessive movements of the upper limbs, active manipulation of the generator, large size pocket, or dementia, are observed, preventive maneuvers like anchoring the generator to the bottom of the pocket or sub-pectoral implants should be considered during implantation. Furthermore, Twiddler's syndrome usually occurs one year or less after implantation of the device [1–15]. In the case presented, the condition was diagnosed 5 months after the implantation. Therefore, we should be aware that Twiddler's syndrome may occur within one year, especially in high-risk cases such as those described above.

In conclusion, we have presented a case of Twiddler's syndrome that was detected by the patient's complaint of generator rotations in the subcutaneous pocket. Typically, Twiddler's syndrome is identified by a sudden device malfunction; however, in this case, the patient's complaint of the generator rotating contributed to an early diagnosis, which prevented the occurrence of a device malfunction.

References

[1] Bayliss CE, Beanlands DS, Baird RJ. The pacemaker-twiddler's syndrome: a new complication of implantable transvenous pacemakers. Can Med Assoc J 1968;99:371–3.

[2] Hill PE. Complications of permanent transvenous cardiac pacing: a 14-year review of all transvenous pacemakers inserted at one community hospital. Pacing Clin Electrophysiol 1987;10:564–70.
[3] de Buitleir Michael, Charles C. Twiddler's syndrome complicating a transvenous defibrillator lead system. Chest 1996;109:1391–4.
[4] Pereira PL, Trubenbach J, Famsworth CT, et al. Pacemaker and defibrillator Twiddler's syndrome. Eur J Radiol 1999;30:67–9.
[5] Gronefeld G, Kleine P, Israel CW, et al. Twiddler syndrome in a subpectorally implanted cardioverter defibrillator. J Cardiovasc Electrophysiol 2002;13:94.
[6] Nicholson WJ, Tuohy KA, Tilkemeier P. Twiddler's syndrome. N Eng J Med 2003;348:1726–7.
[7] Fahraeus T, Hoijer CJ. Early pacemaker Twiddler's syndrome. Europace 2003;5:279–81.
[8] Sholten MF, Thronton AS, Theuns DA, et al. Twiddler's syndrome detected by home monitoring device. PACE 2004;27:1151–2.
[9] Maurizio G, Francois R, Carlo C, et al. Hiccups and dysphonic metallic voice: a unique presentation of Twiddler syndrome. Circulation 2006;114:534–5.
[10] Veltmann C, Borggrefe M, Schimpf R, et al. Fatal inappropriate ICD shock. J Cardiovasc Electrophysiol 2007;18:326–8.
[11] Atar I, Acil T, Ozin B. Reel syndrome in a patient with a three-chamber implantable cardioverter-defibrillator. Europace 2007;9:674.
[12] Spencker S, Poppelbaum A, Muller D. An unusual cause of oversensing leading to inappropriate ICD discharges. Interventional J Cardiol 2008;129:e24–6.
[13] Aleksandra C, Ewa M, Piotr K. Twiddler's syndrome in a patient with a single-chamber implantable cardioverter defibrillator. Europace 2008;10:366.
[14] Gungor H, Duygu H, Yildiz BS, et al. Twiddler's syndrome as a rare cause of implantable cardioverter defibrillator malfunction. J Cardiovasc Med 2009;10:352–3.
[15] Daniel P. Iatrogenic Twiddler's Syndrome. J Interventional Card Electrophysiol 2010;29:135–7.